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## Hemangiopericytoma/solitary fibrous tumor of pectoralis major muscle mimicking a breast mass

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## ABSTRACT

**INTRODUCTION:** Hemangiopericytoma (HPC)/solitary fibrous tumor (SFT) is a very uncommon tumor of uncertain malignant behavior. In 1942, Stout and Murray first characterized these neoplasms as “vascular tumors arising from Zimmerman’s pericytes” and till now hemangiopericytomas and solitary fibrous tumors of the soft tissues are regarded as features of the same entity in the soft tissue fascicle.

**PRESENTATION OF CASE:** We present a case of hemangiopericytoma/solitary fibrous tumor of the pectoralis major muscle in a 33-year-old female. She first noticed a painless mass in her right breast. Ultrasound of the breast revealed a large heterogeneously hypoechoic lesion within the pectoralis major muscle. Fine needle aspiration of the tumor did not produce any meaningful result. The lesion was completely removed by surgical resection. Histologically, the tumor had staghorn-like vasculature and immunohistochemistry for CD34 was positive, whereas desmin, smooth-muscle actin, S-100 protein, cytokeratins (AE1/AE3) and epithelial membrane antigen (EMA) were all negative. A diagnosis of hemangiopericytoma/solitary fibrous tumor was rendered.

**DISCUSSION:** Tumors comprising the HPC/SFT spectrum represent a small subset of soft tissue sarcomas and are found virtually at any site in the body. Wide surgical resection can achieve favorable long-term survival.

**CONCLUSION:** Due to the rarity and unpredictable biological potential of these tumors, long-term follow-up is mandatory even after radical resection, because recurrence or development of metastasis may be delayed many years.

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## 1. Introduction

A wide range of soft tissue neoplasms has to be considered in the differential diagnosis of pectoral area nodules. In soft tissues, the histological growth pattern cannot securely establish a correct diagnosis, as many tumors such as myofibroblastoma, schwannoma, synovial sarcoma, dermatofibrosarcoma protuberans, spindle cell carcinoma, leiomyosarcoma, low-grade fibromyxoid sarcoma, malignant peripheral nerve sheath tumor and liposarcoma share similar clinicopathological features. Therefore, it is essential an experienced soft tissue pathologist should evaluate the specimens.<sup>1</sup>

Hemangiopericytoma represents less than 2% of these soft tissue tumors and was once regarded as a vascular, pericyte-derived tumor, but recently has been categorized as a fibroblastic neoplasm similar to solitary fibrous tumor. Both of them probably belong to a

spectrum of a single pathologic entity, a concept supported by their overlapping histological and clinical features.<sup>1,2</sup>

## 2. Presentation of case

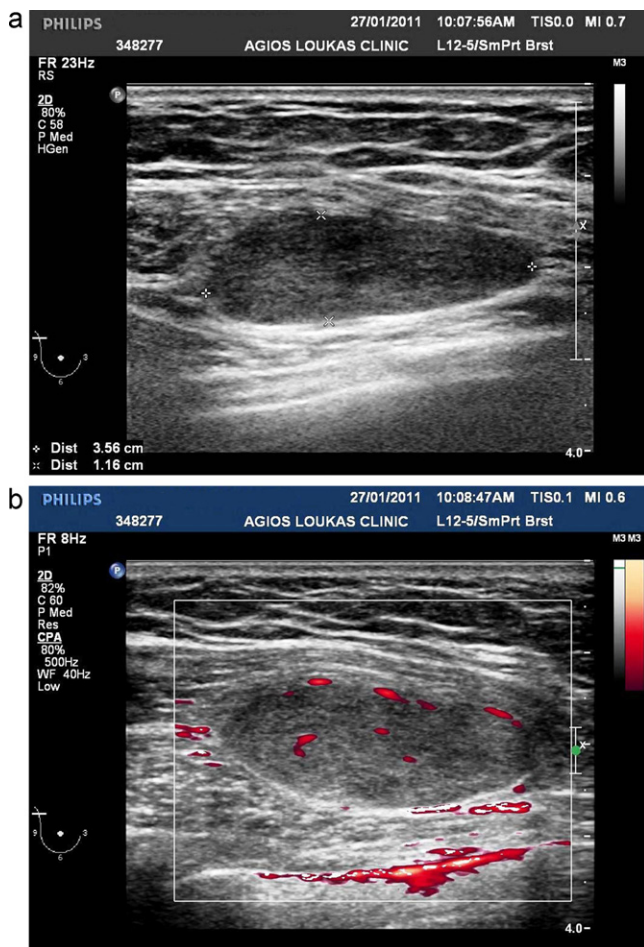
A 33-year-old woman became aware of a non-tender lump in the outer upper quadrant of her right breast. She had first noticed the mass 1 month ago. Her family history was unremarkable, as was her medical history. On physical examination, the patient had a palpable, soft tumor, approximately 4 cm in diameter and was located on the upper outer aspect of the right axillary tail near the chest wall. It was a well-defined, mobile mass, not adherent to either the skin or chest wall. No axillary lymph nodes were found and all laboratory studies were within normal limits.

Targeted breast ultrasonography demonstrated a large heterogeneous and hypoechoic lesion, contiguous to the pectoralis major muscle with its largest dimension approximately 3.5 cm (Fig. 1a). Profound color flow signals were evident in both central and peripheral regions of the mass (Fig. 1b). Fine-needle aspiration was performed, but did not retrieve any meaningful tissue. The patient eventually underwent an excisional biopsy and frozen section

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**Fig. 1.** (a) Targeted breast ultrasonography revealing a heterogeneous hypoechoic lesion confined to the pectoralis major muscle. (b) Increased intratumoral and peritumoral blood flow on Doppler sonography.

analysis confirmed the presence of a mesenchymal tumor of unknown biological potential.

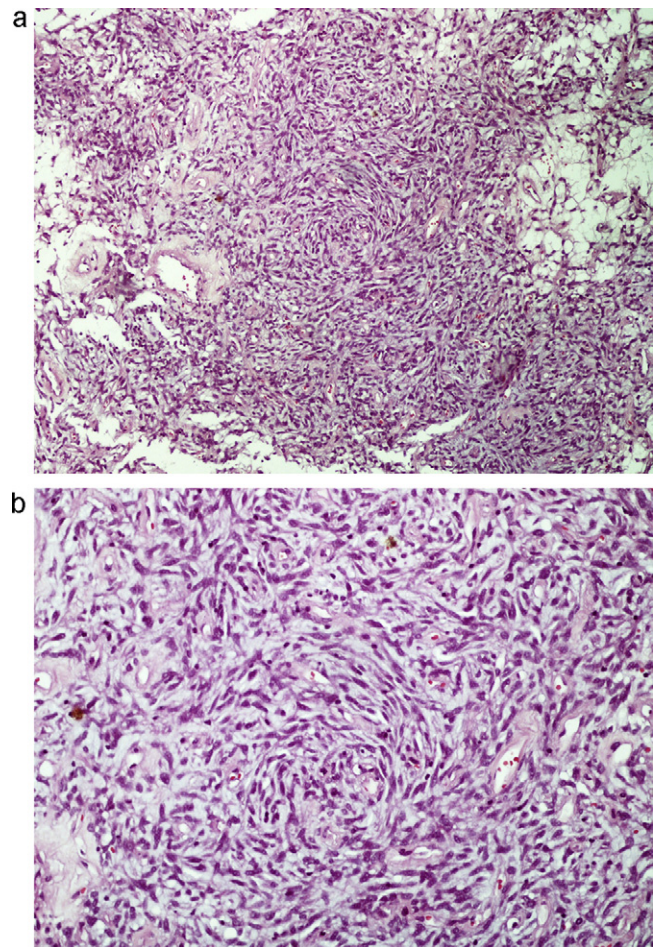
Histology revealed a 5 cm × 3 cm whitish lesion, with a smooth surface, surrounded by a thin, fibrous capsule. The mass was confined to the superficial layer of the pectoralis major muscle, but did not infiltrate the subcutaneous tissue or the pectoral fascia. At higher magnification, there were bundles of spindle and ovoid cells, with moderately irregular nuclei and low mitotic activity (2–3 per 10 high-power fields), resembling a benign mesenchymal tumor (Fig. 2a). Numerous thin-walled and branching vessels within the cytoplasm were present (Fig. 2b). Foci of chronic inflammation, myxoid change, and mast cells were also observed, but no necrosis or cellular atypia was evident.

The immunohistochemical profile included intense membrane positivity for CD34 (Fig. 3), but the lesion did not stain for desmin, smooth-muscle actin (SMA), and S-100 protein (Fig. 4a–c). It was also uniformly negative for cytokeratins (AE1/AE3), epithelial membrane antigen (EMA), whereas the proliferative index was low (Ki-67 < 4%). These diverse histological and immunohistochemical findings therefore established the diagnosis of hemangiopericytoma/solitary fibrous tumor.

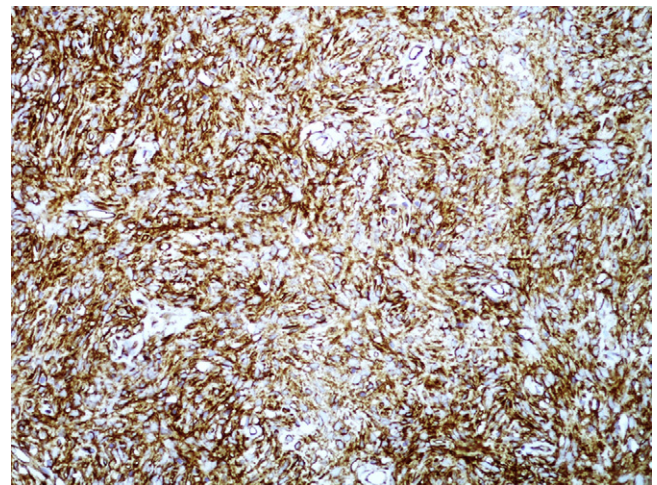
Twenty months later she remains well, without tumor recurrence.

### 3. Discussion

HPC/SFT is an exceedingly obscure tumor of uncertain malignant potential and controversial categorization. Approximately 300



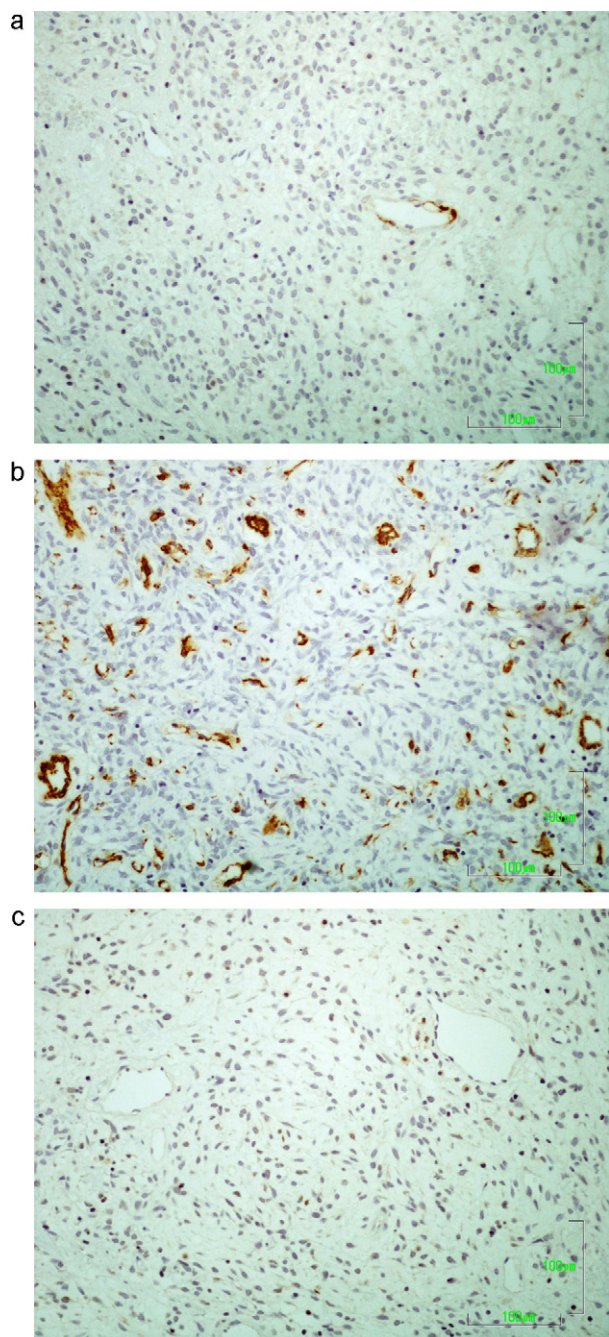
**Fig. 2.** (a) Histologic section showing spindle and ovoid cells, with moderately irregular, without atypia (Hematoxylin and Eosin stain; original magnification ×40). (b) Higher magnification microphotograph demonstrating the presence of thin branching vessels within the mass (Hematoxylin and Eosin stain; original magnification ×100).



**Fig. 3.** Diffuse, strong immunohistochemical staining for CD34 of the spindle-cell component.

cases of HPC have been published since Stout and Murray described HPC as “vascular tumor arising from Zimmerman’s pericytes” in 1942. After further characterization, the WHO reclassified HPC as a fibroblastic/myofibroblastic neoplasm. On the other hand,





**Fig. 4.** Negative expression of the neoplastic cells by (a) desmin, (b) smooth muscle actin, and (c) S-100 protein (Avidin Biotin  $\times 200$ ).

SFT was first reported by Wagner in 1870, as a similar lesion developing from the small oval or spindle-shaped pericytes lining capillaries.<sup>1,3,4</sup>

HPC/SFTs have been described at almost every anatomic location. Most case series showed almost equal distribution for male and female patients, with ages ranging from the third to the eighth or ninth decade with a maximum incidence in the fifth to sixth decade.<sup>4,5</sup> The differential diagnosis is the most demanding and rather confusing issue of a HPC/SFT lesion. Morphologically, HPC/SFT is generally characterized by spindle cell proliferation showing a pattern-less architecture and staghorn vasculature, but the final diagnosis is confirmed by immunohistochemistry using a monoclonal antibody against the human hematopoietic progenitor cell antigen stain (CD34), as in our case report. CD34

immunoreactivity has been reportedly revealed to be strongly and diffusely expressed in many cases of HPC/SFTs, but it is not specific for SFT or HPC alone.<sup>4</sup> Some studies suggest that additional immunoreactivities of bcl2 and CD99 are also diffusely positive in most SFTs. This feature can sometimes differentiate SFTs from HPC, because this spectrum of tumors shares similar histological pattern and CD34 reactivity. However, bcl2 and CD99 were not available in our immunochemistry panel. Vimentin, keratin, smooth muscle antigen (SMA), EMA, desmin, CD117 and S-100 protein are sometimes useful for differential diagnosis of HPC/SFT from tumors with muscle, epithelial or neural origin. Several researchers have proposed that these two neoplasms represent the same entity, especially in lipomatous hemangiopericytoma and the fat-containing variant of SFT. On the other hand, other authors stated that the presence of spindle cell cytological features, various growth patterns with overlapping hypocellular and hypercellular areas and abundant collagenization is indicative of SFT diagnosis, whereas the existence of round or oval nuclei and diffuse stag-horn vascular pattern favors a HPC diagnosis.<sup>6,7</sup> In our case report, HPC/SFT was regarded as the final pathology result, as abundant spindle and ovoid cellularity and widespread stag-horn vasculature were both equally present.

Nuclear atypia, areas of high cellularity or necrosis, increased mitotic index ( $>4$  mitoses per 10 high-power fields), large tumor size ( $>50$  mm) and infiltrative margins are regarded as criteria of HPC/SFT malignancy and positively correlated with local recurrence and metastatic disease. Some studies demonstrated a very low rate of recurrence and distant metastasis, whereas other investigators indicated a relatively increased relapse rate with extended follow-up periods. Therefore, complete resection at an early stage should be the main goal of surgical treatment and follow-up should be maintained for more than 10 years.<sup>1,3</sup> Vallat-Decouvelaere et al. in their study found local recurrence in 4.3% and metastasis in 6.7%, while Gold et al. reported rates of 5.4% and 5.3%, respectively. Tumor relapse occurred after up to 168 months, but most local recurrences or metastases were diagnosed within the first 2 years after surgery. Sites of distant metastasis were lung, liver, bones, mesentery, omentum, mediastinum and retroperitoneum with preference for lung and liver.<sup>8,9</sup>

Nowadays, evolving therapies inhibiting specific angiogenic pathways show promising activities in HPC/SFT. The rich vascular features of HPC/SFT have been long recognized, but the effective value of the vascular endothelial growth factor (VEGF) and its receptor (VEGFR) in hemangiopericytoma/solitary fibrous tumor (HPC/SFT) still remains indistinct. Several phase II trials have investigated the efficacy of bevacizumab, an anti-VEGF antibody, and VEGFR tyrosine kinase inhibitors (TKIs), such as sunitinib, sorafenib and pazopanib, whereas combination therapy with temozolomide and bevacizumab has recently emerged as an encouraging therapeutic regime for HPC/SFT. These targeted agents do not clearly benefit the majority of patients, indicating that biological mechanisms underlying the activity of these agents in HPC/SFT are still poorly understood.<sup>3</sup>

Complete surgical resection is commonly accepted as the current cornerstone of curative intent. Due to improved techniques in reconstructive surgery, large lesions or even recurrent tumors can usually be entirely resected. For HPC/SFT displaying malignant behavior or positive surgical margins after excision, adjuvant radiotherapy is recommended, when feasible. On the other hand, there is no strong evidence supporting the use of adjuvant chemotherapy. Long-term follow is mandatory, because local or distant relapse can occur after resection of an HPC/SFT which appears histologically benign.<sup>1,10</sup>

#### Conflict of interest

There are no conflicts of interest of any kind.

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## Ethical approval

Written informed consent was obtained from the patient for publication of this case report and accompanying images. A copy of the written consent is available for review by the Editor-in-Chief of this journal on request.

## Author contributions

Dimitrios Dragoumis was responsible for original conception and design, editing, English editing, search of the literature, correction, editorship of the manuscript. Klearchos Desiris was responsible for acquisition, analysis and interpretation of data, English editing and search of the literature. Aikaterini Kyropoulou and Anthoula Assimaki were responsible for the histology consulting and pathology examination. Maria Malandri provided ultrasound images and contributed to correction and search of the literature. Aris Tsiftoglou was responsible for correction, editing, revision, and approval of the final version.

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